Myxomatous degeneration of mitral valve *M-mode and two dimensional echocardiographic findings*

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SUMMARY This report presents a patient with an unusual clinical course associated with a mitral "mass" recorded by M-mode and cross-sectional echocardiography. The "mass" was confirmed at operation to be redundant myxomatous mitral valve leaflets. The problem of echocardiographic resolution in the setting of mitral valve prolapse and of the differential diagnosis of a mitral mass is discussed in detail.

M-mode and two-dimensional echocardiography are extremely useful in showing normal and abnormal mitral valve motion.¹ We report an unusual case of a patient with a mitral "mass" documented by M-mode and two-dimensional echocardiography, presenting with possible emboli from the mitral valve, who was found at surgery to have grossly flail and redundant anterior and posterior mitral valve leaflets. The importance of recognising that mitral valve prolapse can progress to flail mitral leaflets and the difficulty of documenting a true mass by M-mode as well as two-dimensional echocardiography in this setting is emphasised. The differential diagnosis is also described in detail.

Case report

The patient was a 55-year-old white man. He was well until 1940 when on physical examination he was noted to have an apical systolic murmur. He was given prophylaxis for bacterial endocarditis. The patient did well until 1964 when he noted the onset of bilateral blurred vision, which he described as "like wearing dirty glasses". The episodes lasted for approximately 45 minutes without neurological sequelae. He did well until he had two more episodes in 1969 and 1973. Ophthalmological and neurological evaluations were negative. In 1976, he had two more episodes of blurred vision—one episode was associated with slurred speech and the other with dizziness. In January 1978, the episodes of blurred vision increased in frequency

to one every two weeks. He was started on acetylsalicylic acid 650 mg twice daily with resolution of his visual disturbance. Further cardiological evaluation with M-mode and two-dimensional echocardiography showed a large mass-like effect on both the anterior and posterior mitral valve leaflets; he was therefore transferred for cardiac catheterisation and consideration for mitral valve replacement. There was no history of rheumatic fever, syncope, orthopnoea, paroxysmal nocturnal dyspnoea, chills, sweats, seizures, and no family history of heart disease.

His blood pressure was 100/82 mmHg. The examination was unremarkable except for the following: the patient had a palpable systolic thrill at the apex, maximal in the left anterior axillary line. There was a grade 4/6 holosystolic murmur at the left sternal border radiating to the axilla, a diastolic sound felt to be either an opening snap or "tumour plop", and a grade 2/6 mid-diastolic rumble at the apex. There were no rubs, clicks, cyanosis, clubbing, or oedema.

Routine laboratory studies including erythrocyte sedimentation rate, Venereal Disease Research Laboratories test, rheumatoid factor, anti-nuclear factor, complement levels, protein electrophoresis, and blood cultures were all within normal limits. The electrocardiogram, vectorcardiogram, and chest x-ray examination demonstrated left ventricular and left atrial enlargement. The M-mode (SKF Ekoline with Irex recorder) and two-dimensional echocardiograms (Varian V 3000) showed multiple thick echoes on the posterior as well as the anterior mitral valve leaflets (Fig. 1A, 1B, 2A, and 2B).

Cardiac catheterisation disclosed a raised pulmonary artery pressure of 36/12 mmHg, with a pulmonary artery wedge V wave of 24 and a mean

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Fig. 1(A)

Fig. 1(B)

Fig. 1(A) *M-mode echo. Continuous strip chart recording with findings of multiple thick shaggy echoes on anterior and posterior mitral valve leaflets.*
(B) *M-mode echo. Continuous strip chart recording with findings of systolic and diastolic fluttering, echoes displaced into left atrium during systole, dilated left atrium, hammock-like posterior bulging of posterior leaflet.*

Fig. 2(A)

Fig. 2(B)

Fig. 2(A)
Two-dimensional
echocardiogram.
Masses or
thickening of
anterior and
posterior leaflets of
mitral valve in this
stop-frame shot.
(B) Two-dimensional
echocardiogram
outline of Fig. 2(A).

pressure of 18 mmHg. The inferior vena cava and right atrial angiograms by hand injection were normal. Pulmonary angiography in the right anterior oblique projection showed a sizeable "mass" in the laevophase on the mitral leaflets moving back and forth into the left atrium, which was severely dilated. Coronary angiography was normal.

On 19 July 1978 during operation the patient was found to have grossly redundant anterior and posterior mitral valve leaflets without ruptured chordae tendineae. There was no evidence of thrombus, tumour, or vegetations on or around the valve. He underwent mitral valve replacement with a No. 29 Björk-Shiley valve. His postoperative course was uncomplicated. On follow-up he has done well without recurrence of his visual disturbance.

Gross and microscopical findings were as follows. The atrial appendage was normal without findings of Aschoff's bodies. The anterior and the posterior leaflets were redundant. The redundant portions were thickened, nodular, and firm with foci of mineralisation. There were histological findings of myxomatous infiltration resulting in disrupted fibrous and elastic elements. Histochemical stains disclosed an increase in acid mucopolysaccharides in the myxomatous tissue. The chordae tendineae were elongated, thin, and not fused. Rare chordal thickening disclosed an increase in acid mucopolysaccharide deposition.

Discussion

M-mode echocardiography enhances the axial evaluation of mitral valve prolapse, valvular vegetations, tumours, and flail mitral valve leaflets.²⁻⁴ Two-dimensional echocardiography adds to the M-mode technique by providing spatial information of the lateral, superior, and inferior cardiac wall motions. Both techniques are limited, however, by gain-setting, resolution, and angulation. When two-dimensional echocardiography is studied during single frame views, there is also degradation of the image and loss of visual integration of motion.¹

Our patient on M-mode echocardiography (Fig. 1A and 1B) seemed to manifest findings characteristic of mitral valvular vegetations as well as fibrosis and calcification. The anterior leaflet was irregularly thickened but less so than the posterior leaflet. There was a coarse mass of thick, shaggy echoes closely associated with the posterior leaflet. Evidence against vegetations was that the posterior leaflet motion appeared restricted and the valvular thickening was uniform. Furthermore, the echocardiographic presentation of a mass, density, or aggregate of echoes posterior to the anterior mitral leaflet, with displacement posteriorly into the left atrium during systole and with abrupt mid-systolic posterior motion, can also mimic a myxoma or thrombus.⁵ Evidence against a myxoma or

thrombus was first that the anterior cusp diastolic closure slope was normal and not decreased, and secondly that the cloud of echoes was not immediately posterior to the anterior mitral valve.

Our patient's echocardiogram also illustrates the confusion and limitations of M-mode echocardiography in patients with mitral valve prolapse.⁶⁻⁹ Chandraratna and Langevin⁶ found that 40% of patients with mitral valve prolapse had shaggy echoes resembling those seen in valvular vegetations. The echoes were thought to be caused by redundant leaflets presenting multiple surfaces to the transducer. The more severe the prolapse, the greater the degree of redundancy and hooding and the more echoes recorded. Without question, noninvasive localisation of a vegetation facilitates expeditious and appropriate medical and/or surgical intervention. Artefactual "vegetations" can be created by high gain settings and can be mimicked by calcification, fibrosis, and mitral valve prolapse (as demonstrated in our patient).

Humphries *et al.*⁴ have reported several M-mode features of flail mitral valve leaflets or ruptured chordae tendineae also found by continuous sector scanning. These authors described paradoxical movement of the anterior and posterior leaflets during diastole, associated hammock-like posterior bulging of the posterior leaflet during systole, severely erratic early diastolic and systolic fluttering of the leaflets, with the anterior leaflet touching the intraventricular septum, abnormal systolic echoes in the left atrium moving toward the posterior left atrial wall, posterior systolic motion of the left atrium, pronounced systolic motion of the left atrium, and distinct systolic motion of the intraventricular septum. Evidence against anterior leaflet rupture in our patient was that there was no increased rate and amplitude of the anterior cusp opening and no coarse diastolic fluttering. Our patient with mitral valve prolapse did, however, present some of the echocardiographic features of a flail mitral valve. If the diagnosis of a flail mitral valve is contemplated, the timing of treatment, specifically surgery, is critical. Therefore, the accuracy of the diagnosis is imperative. Other disorders with features resembling a flail mitral valve (parachute mitral valve, Capetown valve, atrial tumour or clot, aortic regurgitation, atrial fibrillation and flutter, ventricular dysfunction, and congenital defects such as endocardial cushion defects) can be distinguished from a flail valve by other echocardiographic findings. Mitral valve prolapse, however, stands out as the exception and can be virtually indistinguishable by echocardiography.

With the advent of two-dimensional echocardiography, there was new hope of additional accuracy in the detection of vegetations, tumours, and flail mitral valve, especially in patients with mitral valve pro-

lapse.^{10,11} Our patient's two-dimensional echocardiogram (Fig. 2A and 2B) shows apparent masses or thickening on both the anterior and posterior mitral valve leaflets prolapsing in real-time into the left atrium. The apical view was specifically selected for its sensitivity in detecting the structural abnormality of the mitral valve and its apparatus. The findings were suggestive of vegetations, tumours, flail leaflets, or mitral valve prolapse. The lack of resolution by the two-dimensional echocardiogram in delineating the multiple folds of both leaflets renders a definitive diagnosis of mitral valve prolapse difficult and indistinguishable from the other entities discussed. Future application of this technique will depend upon improving its resolution and studying its sensitivity and specificity in a large population of patients with suspected mitral valve prolapse.

Although the absolute diagnostic criteria for mitral valve prolapse have not been established, the left ventricular angiogram has been considered to be the most accurate laboratory test available, though it has limitations, especially with anterior leaflet prolapse. Our patient's angiogram in the right anterior oblique projection showed an abnormal mass or bulge moving backward into the left atrium and forward into the left ventricle. The mass or bulge projected not only anterolaterally but also anterosuperiorly, suggesting involvement of both leaflets. There was no cleft or radiolucency seen between the anterior and posterior leaflets nor any temporal dissociation between the leaflets. Furthermore, differentiation of a mass from prolapse of both leaflets by angiography was difficult for two reasons: (1) a pulmonary angiogram was done rather than a left ventricular angiogram for fear of dislodging a portion of the mass. This resulted in less contrast than necessary to visualise combined leaflet prolapse, and (2) the significant mitral regurgitation further obscured delineation of the valvular involvement.

The association of cerebral ischaemic events and prolapsing mitral valve is receiving more attention as we learn more about the natural history of mitral valve prolapse. The aetiology of our patient's transient ischaemic attacks remains unexplained though his favourable response to acetylsalicylic acid suggests a platelet embolic source, possibly emanating from his abnormal mitral valve. The gross and histological examination of his mitral valve, however, failed to show significant thrombus formation. We concur with the recommendations of Barnett *et al.*¹² that either platelet inhibitor treatment (aspirin, dipyridamole, or sulphinpyrazone) or warfarin-type anticoagulation seems to be indicated in these cases of cerebral emboli in which bacterial endocarditis has been excluded by appropriate cultures.

In summary, our patient illustrates the role of

two-dimensional echocardiography versus M-mode recording in evaluating patients with mitral valve prolapse. M-mode echoes are extremely dependent on transducer angulation and resolution. Two-dimensional echoes add a spatial perspective but are also hampered by spurious echoes, dropout echoes, resolution difficulties, and overlapping echoes from transducer position.¹³

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